

Takayasu Arteritis with Multiple Intracranial Aneurysms A Case Report

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Key words: Takayasu arteritis, multiple intracranial aneurysms

Summary

A rare case of multiple cerebral aneurysms associated with Takayasu aortitis is reported. Only seven cases have been reported so far, all of which are from Japan. This is the first case with this association being reported from India.

A 50-year-old hypertensive woman developed sudden onset severe headache and loss of consciousness. CT scan revealed subarachnoid haemorrhage in the suprasellar cisterns and mild hydrocephalus. The patient on admission had a rebleed producing a massive subarachnoid haemorrhage. Angiogram revealed occlusion of bilateral vertebral arteries and right common carotid artery. The left carotid artery was ectatic with saccular aneurysms in the ophthalmic segment and communicating segment.

Aortogram showed the radiological picture of Takayasu arteritis Type III. Surgical clipping of the aneurysms was performed. However, the patient developed a massive right cerebral infarct in the postoperative period following which she expired. Abnormal haemodynamics in the overloaded cerebral circulation rather than primary arteritic involvement of intracranial vessels appears to be the cause of aneurysm formation.

Introduction

Takayasu arteritis is an inflammatory disease of unknown cause affecting the aorta and its

main branches and is found mainly in young women. It was first reported in 1908 by Takayasu, a Japanese ophthalmologist¹. Cerebrovascular involvement in the form of stenosis of the proximal cerebral arteries has been reported in as many as 24% cases of aortoarteritis². Isolated case reports of intracranial aneurysms associated with Takayasu arteritis have only been reported from Japan. This is the first case of Takayasu arteritis with multiple intracranial aneurysms being reported outside Japan. The presence of hypertension in almost all the reported cases indicates that abnormal intracranial haemodynamic forces play a definite role in the development of aneurysms.

Case Report

A 50-year-old postmenopausal woman, a known hypertensive for the last five years and on irregular medical treatment, had an episode of sudden onset severe occipital headache associated with projectile vomiting and loss of consciousness. Following the episode she developed bilateral diplopia on lateral gaze. The patient had a history of headache, malaise and intermittent claudication in the right upper limb for the last eight years. The patient had bilateral lateral rectus palsy on neurological examination and absent pulses in the right upper limb. A CT study of the head revealed subarachnoid



Figure 1 CT head study shows subarachnoid bleed involving suprasellar and right-sided ambient cistern with hydrocephalus.

haemorrhage in the suprasellar cisterns and mild hydrocephalus (figure 1). Laboratory investigations showed erythrocyte sedimentation rate (ESR) of 41mm/h. The patient was admit-

ted to the ward where she had a sudden episode of unresponsiveness and acute respiratory arrest, following which she was intubated and put on ventilatory support. A repeat CT study showed evidence of rebleed with massive subarachnoid haemorrhage involving the supratentorial cisterns and the ventricular system.

An emergency cerebral angiogram and aortogram revealed occlusion of bilateral subclavian artery origins and the right common carotid artery from its origin up to a point just proximal to its bifurcation (figure 2). The sole artery supplying the intracranial circulation was the left common carotid artery which was dolichoectatic and showed saccular aneurysms arising from the ophthalmic and communicating segments of the internal carotid artery (ICA).

The posterior communicating artery (PCOM) was also ectatic and tortuous (figure 3). The aortogram study showed post subclavian narrowing of the thoracic aorta, mild left renal artery origin stenosis, celiac and superior mesenteric origin stenosis and infrarenal aortic narrowing consistent with radiological features of a type III Takayasu arteritis (figure 4).

Surgical clipping of the aneurysms was attempted. The patient developed a massive right-sided cerebral infarct in the immediate postoperative period and expired on the third postoperative day.

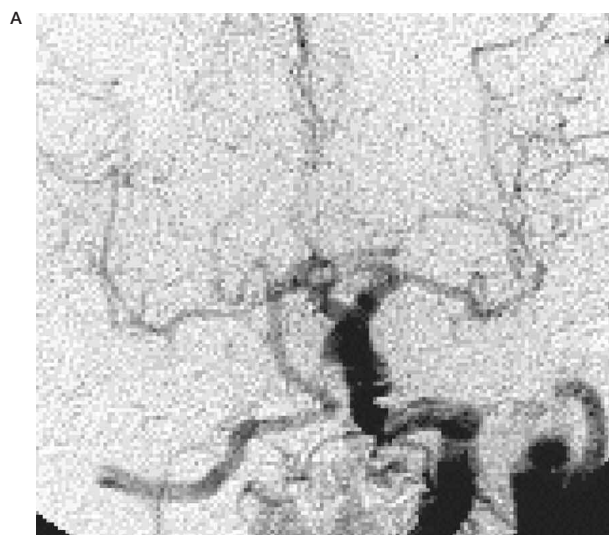


Figure 2 Left common carotid study AP and lateral views showing saccular aneurysms in the communicating and ophthalmic segments of the left ICA and ectatic left PCOM.

Figure 3 Arch aortogram study showing occlusion of the brachiocephalic and left subclavian with post subclavian severe stenosis of the aorta. Delayed image shows retrograde filling of both subclavian arteries via vertebrals.



Discussion

Takayasu arteritis is predominantly a disease of young females, in their second and third decade^{3,4}. However, the course of Takayasu arteritis extends over many years with cases presenting with symptoms secondary to arterial insufficiency much later in life⁵. The distribution of the disease is more common in Japan and the Indian subcontinent, but studies have shown an increasing incidence in Mexico, Brazil and Columbia^{6,7}. The disease was classified into four different types according to its topographical distribution by Lupi-Herrera³. Type I shows aortic arch and its branch involvement whereas type II shows involvement of the thoracoabdominal aorta and its branches. The present case falls into type III with features of both type I and type II. The type IV disease involves the pulmonary arteries in addition to any of the above types. A type V has been proposed showing additional involvement of coronary ostia⁸.

The disease has an active stage characterized by systemic symptoms, diffuse granulomatous inflammation involving the vessel wall on histopathology and a chronic healed stage characterized by fibrotic changes in the vessel wall on histopathology. Persistently elevated erythrocyte sedimentation rate (> 40 mm) was identified as a reliable indicator for activity of inflammatory process and disease progression⁹. Cerebrovascular involvement have been reported in as many as 24% cases of aortoarteri-

tis with occlusion, stenosis, aneurysms and arterial wall thickening seen mostly in the extracranial carotid and vertebral circulation². The incidence of aneurysms in Takayasu arteritis involving the aorta and its major branches is as high as 31.9%¹⁰. An association between aneurysm formation and systemic hypertension in Takayasu's arteritis has also been proposed suggesting aggressive treatment of hypertension¹¹. Intracranial aneurysms associated with

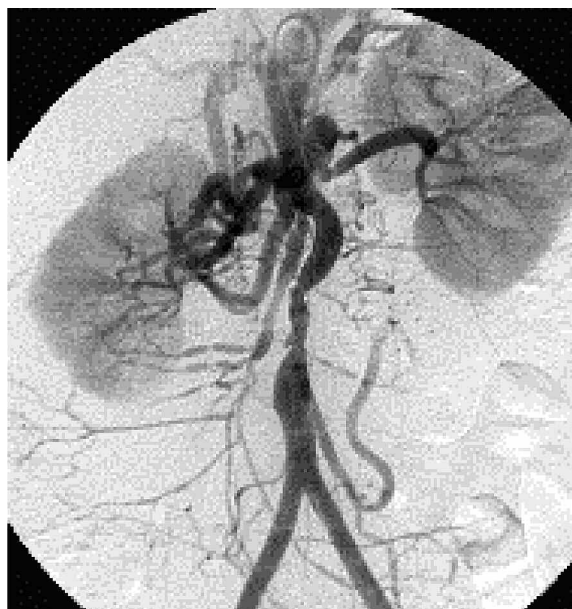


Figure 4 Abdominal aortogram study showing infrarenal aortic, left renal artery and SMA origin stenosis.

Takayasu arteritis are a rare complication reported in seven Japanese patients to date¹². The predominant site of involvement was the basilar bifurcation with multiple aneurysms seen in only one case¹³.

The present case is the only one described outside Japan and only the second case with multiple intracranial aneurysms. The presence of dolichoectasia in the sole artery supplying the brain in the present case indicates the role of haemodynamic factors in the pathogenesis of intracranial aneurysms rather than primary

involvement of intracranial vessels by the arteritic process. The presence of hypertension in our cases as well as the majority of previously reported cases also suggests a role for haemodynamic factors as the cause of aneurysm. Aggressive treatment of hypertension in Takayasu aortitis as stressed by Sharma et al is therefore important in decreasing the risk of aneurysm formation¹¹. Aneurysm formation and subarachnoid haemorrhage must also be considered along with ischaemic changes as neurological complications of Takayasu aortitis.

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EDITORIAL COMMENT

The authors report a very interesting aneurysm case as they are quite rare in this particular disease at least intracranially. One paper published in 1989 already pointed to such an association¹. One can think that the frequency of the intracranial complications of Takayasu's disease will probably be modified in the future with the use of non invasive exploration like angio-MR. Yet this possibility remains probably insufficient as reported in a recent article² with two cases of subarachnoid haemorrhage in Takayasu patients in which exploration failed to demonstrate aneurysms.

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